

## Case report

# Coronary arteritis complicating rheumatoid arthritis

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**SUMMARY** The case is described of a patient with rheumatoid arthritis (RA) who developed heart failure and vasculitis and died of a myocardial infarction. Autopsy showed vasculitis of several major epicardial coronary arteries. Coronary arteritis in a patient with RA is seldom diagnosed during their lifetime but should be suspected when such a patient develops ischaemic heart disease.

**Key words:** vasculitis, myocardial infarction.

Rheumatoid arthritis (RA) is a joint disease that can be complicated by extra-articular manifestations such as rheumatoid nodules, episcleritis, neuropathy, serositis, nailfold lesions, petechiae or palpable purpura of the skin, and skin ulcers. Several of these manifestations are supposed to originate from vasculitis.<sup>1–3</sup> Histological proof for the clinical diagnosis is most easily found in biopsies of the skin, muscle, or rectal mucosa.<sup>4</sup>

Systemic vasculitis complicating RA has a reported mortality rate of up to 30%.<sup>5–7</sup> It has been demonstrated in necropsy studies that vasculitis in rheumatoid arthritis can also affect the coronary arteries, mostly of the smaller muscular type.<sup>8,9</sup> Until now only three patients have been described who, during their lifetime, were suspected of having coronary arteritis.<sup>4,8,9</sup> We report a patient with rheumatoid arthritis who developed heart failure and vasculitis of the skin and died of a myocardial infarction due to vasculitis of several major coronary vessels.

## Case report

A male patient, born in 1914, with seropositive RA since 1960, was admitted in April 1985 because of malaise, weight loss, and severe joint pain. Previously he had been treated with non-steroidal

anti-inflammatory drugs, chloroquine, gold, D-penicillamine, and azathioprine.

In May 1983 the patient developed multiple petechiae. A skin biopsy showed leucocytoclastic vasculitis. Because an allergic reaction to azathioprine could not be excluded this medication was stopped. The patient was then treated with prednisone 10 mg and indomethacin 150 mg daily. The skin of the lower legs continued to show petechiae and palpable purpura. An electrocardiograph (ECG) in 1984 (during an admission for evaluation of disease exacerbation) showed sinus rhythm and a mild disturbance in intraventricular conduction. The ST segments were normal.

In February 1985 he was admitted with congestive cardiac failure and the ECG showed flattened, but not depressed, ST segments in II, III, AVF, V5, and V6. After treatment with frusemide and digoxin the T waves became inverted in leads V1–4, compatible with a digoxin effect.

The results of physical examination on admission were: body weight 50 kg (63 kg one year before), blood pressure 100/60 mmHg, pulse rate 80/min, temperature normal. There was episcleritis of the right eye. A few rales were heard over the basal areas of the lungs. No heart murmurs or gallop sounds were heard. Liver and spleen were not palpable and there was slight pitting oedema around the ankles. Most joints showed severe limitations of movement and there were signs of an active synovitis of the right knee. Periosteal nodules were palpable on the dorsum of both elbows. The skin of

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the lower legs showed multiple petechiae and palpable purpura.

Selected laboratory investigations gave the following results: erythrocyte sedimentation rate 105 mm/1st h, haemoglobin 5.8 mmol/l (93.4 g/l), hypochromic, microcytic, white blood cell count  $7.1 \times 10^9/l$  with 34% band forms and 2% eosinophils, platelet count  $561 \times 10^9/l$ , urea 4.4 mmol/l, creatinine 51  $\mu\text{mol/l}$ , calculated creatinine clearance 70 ml/min, creatine phosphokinase 11 EU (normal 5–50).

Total protein was 68 g/l, albumin 29 g/l, no paraprotein demonstrable. Rose-Waaler test 400 IU, latex fixation 400 IU, antinuclear antibodies strongly positive, anti-deoxyribonucleic acid antibodies negative. Circulating immune complexes as measured by the C1q binding assay were 2242  $\mu\text{mol}$  aggregated IgG/ml (normal <10). Total haemolytic complement 42 U/ml (normal 256–580), C4 14 mg/dl (normal 17–30), C3 60 mg/dl (normal 68–104). The urine gave a negative test for protein, and the sediment contained no abnormalities. Three blood cultures were negative. Tests for occult blood in the faeces were repeatedly negative.

The electrocardiogram showed first degree atrioventricular block (pr: 0.22 s) and a configuration compatible with a right bundle branch block in lead V1. There was also evidence of a left anterior hemiblock with a left axis deviation. Echocardiography showed an enlarged left atrium and ventricle and dyskinetic motion of the lateral left ventricular wall. A chest radiograph disclosed an enlarged left ventricle of the heart, no signs of pulmonary vascular engorgement, and a slight pleural effusion on both sides.

A biopsy of the affected skin showed leucocytoclastic vasculitis and perivascular deposits of IgM and

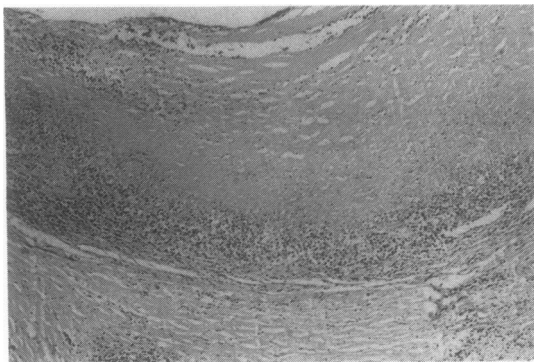


Fig. 1 Coronary artery showing thickening of intima and dense infiltration of inflammatory cells, particularly in the media (haematoxylin and eosin).

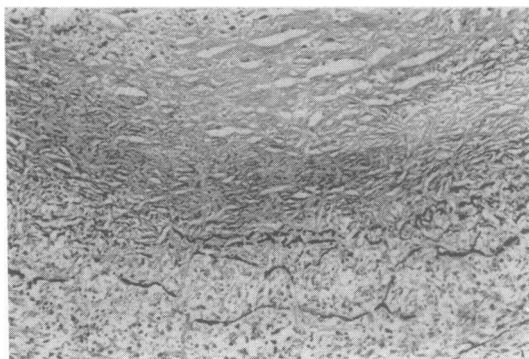


Fig. 2 Coronary vessel wall with fragmentation of the inner elastic membrane in an area of inflammation (Elastica van Gieson).

C3. The diagnosis was rheumatoid arthritis complicated by systemic vasculitis and cardiomyopathy either due to atherosclerosis or vasculitis. Treatment with high doses of corticosteroids and cyclophosphamide was considered but refused by the patient. On the sixth day of admission the patient was suddenly found dead.

Postmortem examination showed several major epicardial coronary arteries (diameter up to 4 mm) with massive infiltration of the media and adventitia by plasma cells, lymphocytes, and histiocytes (but no giant cells), accompanied by proliferation of capillaries (Fig. 1). This infiltrate led to obscuring and destruction of the muscle cells of the media and interruption of the internal elastic membrane (Fig. 2). The intimal layers of these arteries showed massive thickening by fibrous tissue proliferation with occasional atheroma and focal calcification and with some infiltrate, but much less than in the media. The lumina were markedly narrowed, but thrombosis was absent.

The anterior part of the left ventricular wall showed a recent infarction and in the lateral part scars were found. Owing to this heart lesion, bilateral pulmonary oedema (lungs together 1500 g) and bilateral pleural effusions (200 ml) were present. In the lungs, larynx, and testicles some small arteries showed fibrous thickening of the intimal layer with lymphocytes in the vessel walls, indicating proliferative vasculitis. The pericardial cavity was obliterated. No signs of amyloidosis were found.

## Discussion

According to the literature there is no increased incidence of myocardial infarction in rheumatoid patients.<sup>10 11</sup> In our Leiden material of 173 autopsies

on patients with RA 17 had a fatal myocardial infarction compared with 20 in control autopsies on subjects without RA (matched for sex, age, and autopsy period). Autopsy studies showed that the pathological process underlying ischaemic heart disease in RA is usually atherosclerosis.<sup>12 13</sup>

Although vasculitis of the coronary arteries is seldom diagnosed during a patient's lifetime,<sup>12 14 15</sup> it is found more frequently at autopsy. A necropsy study of 100 patients with RA showed 'patchy' arteritis of the coronary vessels to occur in 20%.<sup>9</sup> This finding was confirmed by Lebowitz.<sup>8</sup> The vasculitis was found most often in the smaller intramyocardial arteries.<sup>1 8 9</sup> In addition to the present patient our series of 173 RA autopsies showed 17 cases of systemic vasculitis, in four of which the (small and intramyocardial) coronary arteries were affected.

In a clinical study of 50 patients with systemic rheumatoid vasculitis only one patient was found to have coronary arteritis at postmortem examination.<sup>5</sup>

That a clinical diagnosis of vasculitis of the coronary arteries in RA is so seldom made, might be explained in two ways. Firstly, not all of these patients develop clinically evident ischaemic heart disease. Eighteen out of 55 patients with rheumatoid vasculitis of the coronary arteries described in post-mortem studies had developed a clinical myocardial infarction.<sup>12</sup> Secondly, clinically, coronary vasculitis cannot be distinguished from the more frequently occurring atherosclerotic heart disease. Arteriography may be of help in establishing the diagnosis.<sup>15</sup>

This patient with longstanding RA developed clinically evident heart failure two months before admission. He was admitted because of active rheumatoid disease with vasculitis and died suddenly of a myocardial infarction due to vasculitis of some major coronary arteries. Since early diagnosis

followed by prompt immunosuppressive therapy has been reported to be very effective in patients with systemic rheumatoid vasculitis,<sup>16</sup> this treatment should be considered when a patient with rheumatoid arthritis and systemic vasculitis develops ischaemic heart disease.

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